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Case Report,

Recurrent Multiple Symmetric Lipomatosis Affecting the Upper & Lower Limbs: A Case Report

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Abstract:
Madelung disease is a rare, metabolic, malignancy-associated and disfiguring condition of unknown etiology, characterized by potentially debilitating neurological, cardiopulmonary, gastric, musculoskeletal and psycho-social complications. Also known as multiple symmetric lipomatosis (MSL), Benign Lipomatosis (BSL), and Launois-Bensaude syndrome, MSL is characterized by the symmetrical growth of subcutaneous non-encapsulated fat deposits, usually around the neck, but in some cases also around shoulders, the upper trunk, chest and limbs, and hip girdle. These abnormal fat deposits may rapidly grow over a few months or slowly enlarge over several years. Expanding fatty deformations may be disabling, have disfiguring aesthetic consequences, and infiltrate or compress adjacent vital structures. This case illustrates that painful, massive growths may also occur around the knees, re-grow post excision, and present with compressive neurological symptoms necessitating further surgical interventions.

Keywords: Madelung, lipomatosis, tumor, metabolic.

Interoduction:

Madelung disease, also known as multiple symmetric lipomatosis (MSL), benign lipomatosis (BSL) or Launois-Bensaude syndrome, is a rare metabolic condition characterized by the growth of massive symmetrical, subcutaneous, non-encapsulated (unlike lipomas) fat deposits on the patient's body, usually around the neck, but in some cases also around shoulders, the upper trunk, chest and limbs, hip girdle. These abnormal fat deposits may grow rapidly over the course of months or more slowly over a period of years.

MSL was first described by Benjamin Brody in 1846. In 1888, Otto Madelung presented 35 cases of this disease, with Launois and Bensaude in 1899, describing another 30 cases of excessive adipose tissue growth around the neck, nape, back and shoulders. This new disease was then named Madelung disease or Launois-Bensaude syndrome. From then to the end of 2002, there were around 300 reported cases of patients with this disease in the medical literature[1, 2]. It should not be confused with Mandelung deformity of the wrist, which is an epiphyseal growth plate disturbance characterized by dorsal and radial bowing of the Radius or Madelung dyschondrosteosis, a dysplasia associated with the Madelung deformity. MSL is also a distinct entity from the similarly named familial multiple lipomatosis. A distinguishing feature is that in MSL, lesions are non- encapsulated. Lipoma size typically increases over time, predisposing to pain and movement limitation. MSL may also progress to liposarcoma[3]. Madelung disease is most commonly seen in the Mediterranean population with a male to female ratio of 15:1. The commonest age of onset is between the third and fifth decades. MSL has also been associated with chronic alcoholism[4], and has been associated with lipid storage diseases. Enzi and colleagues also described two types of lipomatosis based on the distribution of fat tissue: type I is characterized by lipomas located in the nape of the neck, the supraclavicular and deltoid regions (Madelung's collar), while in Type II lipomatosis, fat tissue diffuses extensively into the subcutaneous fat layer giving the patient and appearance 'pseudoathletic' appearance. Type III is a rare thigh girth gynecoid variant with a preponderance of female Type II and III[5]. Aside from disfiguring aesthetic effects,

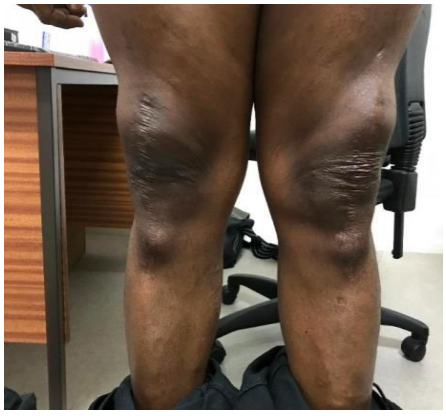
some lipomatous masses may infiltrate adjacent structures; provoke dyspnea, dysphonia, dysphagia, due to compression-infiltration-obstruction of the upper airways and digestive tract. Lesions may also affect neck mobility, and progress to mediastinal involvement and superior vena cava syndrome[6].

Causation theories of metabolic lipid enzyme (catecholamine-induced lipolysis) disturbance, mitochondrial DNA anomalies, and brown fat precursor defects have been proposed[7]. MSL treatments include surgical lipoma removal, liposuction, and avoidance of alcohol consumption[8,3]. However, injection lipolysis therapy may predispose to soft tissue adhesions which present challenges to surgical resection[9].

Case Report:

A 48 year old, right handed male office worker of African descent and originally from southern Nigerian, reported to an outpatient clinic with a history of multiple, painful joint lumps involving the elbows and dorsal fingers and right wrist, which were beginning to interfere with his ability to work. He previously had been a carer but had retrained to work in the business support sector. He had first noted the lumps at age 29. There was no history of prior trauma. He recollected excision of the elbow lump excision 21 years prior, and bilateral knee lump excision some 13 years prior. He was concerned as the elbow, and finger lumps had reoccurred and were starting to affect joint movement and dexterity. Resting his elbows on his desk to type was a problem and accidently bumping his elbows on hard objects would reproduce transient excruciating pain. The dorsal finger lumps had progressively enlarged over the Previous 2 years and were also causing index, middle and ring fingertip numbness and dorsal right greater than left hand night pain.

He also described bilaterally intermittent volar forearm pain, which would sometimes last the entire day. His grip strength was reportedly okay, and he obtained some relief with the application of 'Vick's Vapor Rub,' but no relief with aspirin. He also reported recent and frequent prior trips to the accident and emergency regarding his upper limb pins & needles. He drank alcohol occasionally, was a non-smoker, and had engaged in bicycling, running and gym in order to keep is weight down. He also used to swim and liked recreational sports. However, the lumps had curtailed his ability to participate. He presented with a history of hypercholesterolemia, hypertension, and ischemic heart disease and had undergone coronary artery stenting 10 years ago. His mother and father were both diabetic. His medications were nicorandil, aspirin, paracetamol, pravastatin, bisoprolol, and ramipril. He was not allergic to any medications. Signed consent for anonymous history and images for educational and publication purposes was obtained. On examination, multiple lipomatous overgrowths were overlying the bilateral elbows, metcarpal phalangeal joints. He also had bilateral anterior knee scarring in keeping with a history of prior lumpectomies. He displayed full range of neck, shoulder, elbow, wrist and hand motion. Multiple upper limp lumps of circa 1-7 cm in size were present bilaterally. Upper limb sensation was intact. There were no upper limb dural tension signs and Spurling's was negative. The upper limbs were neurovascularly intact and there were no gross signs of hypermobility.



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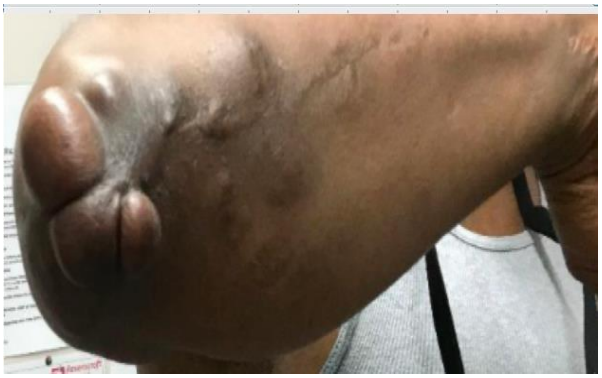

5.

Figure1-5: Post-surgical upper & lower limb reoccurrence of MSL lesions.

Discussion:

MSL is considered to be a rare, benign condition. However, its prevalence, progression to liposarcoma, inherited predisposition and predilection for reoccurrence may be underestimated. The association of MSL to risk factors (i.e. alcohol, smoking) also remains unclear[10]. As this case illustrates, MLS may atypically occur in a non-Mediterranean, non-alcoholic patient. This may also be the first case documenting involvement of the knees. This case also illustrates that massive, neural compressive, non-encapsulated, soft tissue overgrowth may reoccur following primary excision, necessitating further surgical interventions. Though only two cases of liposarcomatous progression of Madelung's disease has been reported in the literature[11,12], progression to malignancy may be underestimated. Biopsy is therefore recommended.

Conclusion:

The patient was referred to orthopaedic surgeons and underwent upper limb surgical resection of the neuro-compressive lesions, with satisfactory abatement of symptoms. Biopsy excluded malignant transformation. Surgical resection is indicated to manage reoccurrence of expanding symptomatic space occupying tumors, which may otherwise have debilitating neurovascular consequence. Liposuction and alcohol abstinence have also been described as managements in the literature[8,3]. Reoccurrence prevention and management strategies include alcohol abstinence. Malignancy surveillance monitoring is a topic of further study.

Conflicts of Interests: None

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